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External Ear Canal Cholesteatoma Occurring 9 Years After Stapes Surgery: A Case Report

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Abstract
While cholesteatoma occurs frequently in the middle ear, external ear canal cholesteatoma (EECC) is a rare entity, which is often misdiagnosed. It is defined as an accumulation of epithelial debris in the ear canal associated to bony erosion. It is divided into primary or spontaneous, and secondary EECCs. The main symptoms are otorrhea, otalgia and hearing loss. Imaging is necessary to confirm diagnosis and to evaluate the disease extension. We report a case of 45-year-old female patient with a history of bilateral stapedectomy for bilateral otosclerosis. After being involved in a traffic accident, she presented hearing loss and tinnitus. Otoscopy showed stenosis of the external auditory canal with accumulated keratin debris and erosion of the posterior wall. The left tympanic membrane was intact but slightly opaque. Pure tone audiometry revealed a mild conductive hearing loss with an averaged air-bone gap of 30dB. Temporal bone HRCT showed, in addition to piston prothesis dislocation, a soft-tissue density lesion in the external auditory canal, with circumferential bony destruction and invasion of the mastoid cells' anterior and superior walls. She underwent surgery for cholesteatoma removal and canaloplasty as well as piston prothesis repositioning, with good outcomes. The particularity of this case is the incidental discovery of EECC, as the patient was asymptomatic until the dislocation of her piston prothesis, which caused her symptoms and prompted her to come to our department, and the concurrent management of an EECC and piston prothesis dislocation.

Keywords: *External ear canal cholesteatoma, hearing loss, stapes surgery, canaloplasty, piston prothesis dislocation*

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I. Introduction

Cholesteatoma is an epidermal inclusion cyst, whose capsule and matrix are formed by stratified squamous epithelium which accumulates desquamated keratin debris concentrically and has the property of bone erosion (1). Johannes Müller introduced the term "cholesteatoma" in 1838 to describe the white-yellow keratin flakes that mimic cholesterol crystals (2). Cholesteatomas are typically located in the middle ear and mastoid, but may also develop in the external auditory canal (EAC). The term “External Ear Canal Cholesteatoma” (EECC) was first used by Scholefield in 1893(3), but the first description of this disease was made by Toynbee in 1850 (4). EECC presents as an accumulation of epithelial debris in the ear canal, and was often confused with keratosis obturans until a clear distinction was made by Piepergerdes et al. in 1980 (5). They reported that EECC is characterized by the invasion of squamous epithelial tissue into localized areas of bony erosion of the EAC, and the presence of osteonecrosis and the formation of bony sequestrum (5). EECC is a rare entity with an estimated incidence rate of 1.2–3.7 per 1000 new otology patients (6,7). EECCs are divided into idiopathic or primary EECCs, and secondary EECCs can be posttraumatic, postsurgical, postinflammatory, postirradiatory, or due to ear canal stenosis (8). Common symptoms include otorrhea, chronic and dull ear pain, hearing loss, ear fullness, and itching (8). In some cases, EECC may be asymptomatic, while in others, it may manifest with complications such as facial palsy (8). Because of its non-specific clinical presentation and rarity, EECC could be misdiagnosed. High-resolution computed tomography (HRCT) is recommended for suspected EECC (9). It shows widening of the EAC with a soft-tissue density mass and, erosion of the walls of the bony ear canal, the extent of lesions and directions of expansion pathways (8,10). While surgical removal is the best option for patients with advanced secondary EECC, patients with small primary EECC can be managed conservatively, with regular debridement under local anesthesia (11). We present a case of incidental discovery of EECC in a 45-year-old woman who had stapes surgery 9 years ago.

II. Case Report

A 45-year-old female patient presented to our department with complaint of left-sided hearing loss associated with ipsilateral tinnitus, ear fullness, otalgia and headaches, beginning since her involvement in a mild traffic accident 3 weeks ago. There was no otorrhea, facial palsy, vertigo. She had a history of bilateral otosclerosis for which she underwent stapedectomy with endaural approach and piston prosthesis placement on the right side in 2014, and on the left side in 2015, with good audiological outcomes. Our first thought was that she had a dislocation of her piston prothesis. The right otoscopy was normal. The left otoscopy revealed a stenosis of the external auditory canal with accumulated keratin debris and erosion of the posterior wall. The left tympanic membrane was intact but slightly opaque. In the attic, there was no obvious keratinized growth. Her cranial nerves examination was normal. In the tuning fork tests, a left conductive hearing loss is assessed as she had Rinne's negative test in the left side and in Weber's test, the sound is heard in the left side. Pure-tone audiometry showed on the left ear, a moderate conductive hearing loss with an averaged air-bone gap of 30 dB. Stapedial reflex was absent. Axial and coronal high-resolution computed tomography (HRCT) sections of the left temporal bone (Figure 1 A, B, and C) showed a soft-tissue density lesion in the external auditory canal, measuring 21.4 mm in transverse diameter, 15 mm in anteroposterior diameter and extending in height over approximately 18 mm, with circumferential bony destruction and invasion of the mastoid cells' anterior and superior walls. The middle ear cleft was unaffected. There was a focal hypodensity of the otic capsule on the oval window's anterior border, and the piston prothesis was dislocated. HRCT on the right side indicated only the otosclerosis hypodensity and the piston prothesis. Diffusion-weighted magnetic resonance imaging (DW-MRI) revealed hyperintense lesion within the left external ear canal, indicating restricted diffusion (Figure 1D). The definite diagnosis of this case was left external ear canal cholesteatoma, that occurred 9 years after a stapes surgery, associated with dislocation of piston prothesis.

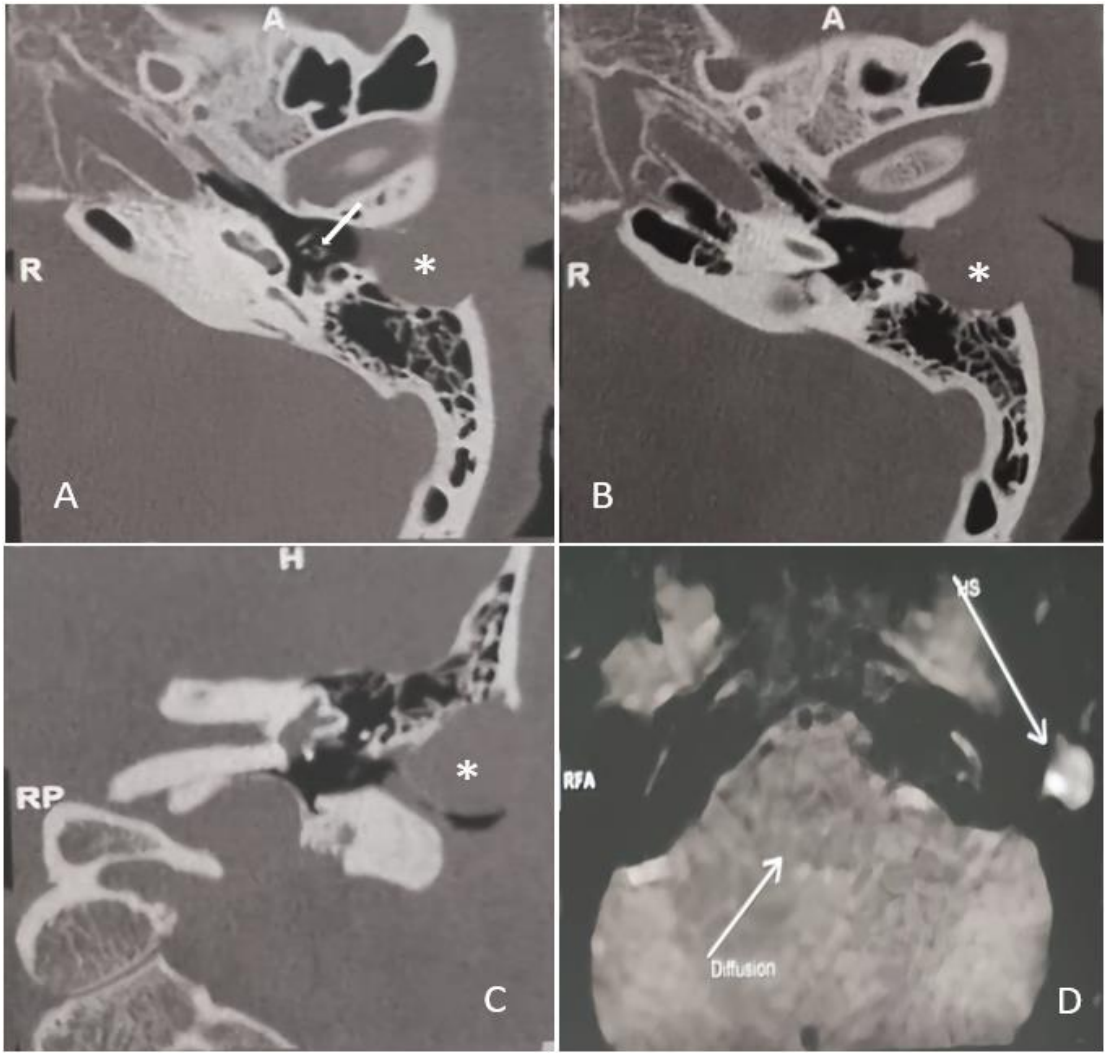

Figure 1*: Imaging of the EECC. A+B: Axial sections of the left temporal bone HRCT showing the soft tissue density lesion in the EAC (*) with invasion of mastoid cells. The white arrow shows the loop of dislocated*

piston prothesis. C: Coronal section of the left temporal bone HRCT showing normal middle ear and soft-tissue density in the EAC (*). D: Diffusion-weighted magnetic resonance imaging (DW-MRI) in axial view showing hyperintense lesion in the left EAC.

As the EECC is classified stage III according to Shin's classification (12), surgery was planned. The patient underwent total cholesteatoma removal using a postauricular approach, modified mastoidectomy, posterior tympanotomy and canaloplasty. The repositioning of a new piston prothesis was done before sealing the opened cavities with concha cartilage and bone dust, and grafting the defect surrounded by healthy skin with fascia and perichondrium. The external ear canal was entirely splinted with silicone sheets, and plugged with Pop-Oto-Wick for 2 weeks. The tissue specimen was sent to histology which confirmed the diagnosis. The post-operative period was uneventful. Oral Amoxicillin with clavulanic acid and topical ofloxacin ear drops were administrated for 10 days. The patient was discharged on 3rd day of surgery. Regular follow up scheduled every month for the first three months, and every three months. The ear canal was well healed by the 2nd month after surgery. No sign of recurrence was found at the 9th month of the follow up.

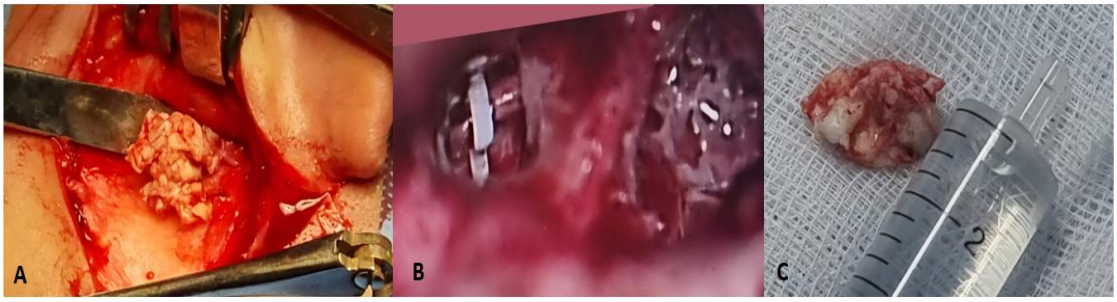

Figure 2 : *Preoperative images. A: EECC after elevation of the tympanomeatal flap via the postaural approach. B : Piston prothesis repositioning. C: EECC specimen sent to histopathological examination*

III. Discussion

External ear canal cholesteatoma (EECC) is a rare disease with an annual incidence rate cholesteatoma in the general population is 0.15 to 0.30 cases per 100 000 people per year (7,8), when compared to 9.2 cases per 100 000 people per year for middle-ear cholesteatoma. EECC is an accumulation of squamous keratinized material in the external ear canal, which causes periostitis and bony erosion (13). According to the expansion pathways, invasion of adjacent structures may be seen, including the mastoid, middle ear cavity and temporomandibular joint, and very rarely the facial nerve canal, labyrinth, sigmoid sinus and skull base (7). EECCs are divided into Primary or idiopathic or spontaneous EECC, and Secondary EECC. The pathogenesis of primary EECC remains unclear, but it may be attributed to microtrauma of external ear canal skin caused by cotton-tipped applicators or hearing aids, epithelial cell entrapment and accumulation due to age-related alterations in epithelial migration and cerumen glands leading to drier wax composition, microangiopathy caused by smoking or diabetes mellitus may compromise the blood supply in the floor of the ear canal, and first branchial arch anomalies (7,14). Secondary EECCs may be categorized into : *Posttraumatic* that results from disrupting the integrity the ear canal skin by all forms of trauma, *Postoperative* in which cholesteatoma develops in the site of a tympanomeatal flap by overlapping or inverting inward of the edges or by entrapment of keratinized epithelium under the graft, *Poststenotic* due to ear canal atresia, exostosis, osteoma, fibrous dysplasia, *Postinflammatory, Postobstructive, Postradiation, and Postumorous* (14,15). Our case is a postoperative secondary EECC, that developed 9 years after stapes surgery. Owen et al. reported a risk of 1.3 cases of EECC per 1000 surgeries representing a variety of procedures (7). Anthony et al. reported a risk of one EECC in 3,000 stapedectomies (6). Secondary EECC is considered to be more common (16), however, primary EECC accounted for 52% of cases in the Owen's study (7) and 34/45 in the Lin's study (13). In addition, He et al. published the largest series of EECC with 301 ears in 279 patients, and all cases are primary (10).

EECC was thought to be most common in older patients, as reported by Piepergerdes et al (5) and Holt who found a mean age of 62 years (15). In more recent studies, younger patients have been diagnosed with EECC, with mean ages of 30 and 46 years in the studies by Dongol (8) and Dhingra (16) respectively. The age of our patient is in the range of recent reports. In the study by He et al, which included 279 patients, the age ranged from 6 to 89 years (10), therefore, EECC may occur at any age. Women appear to be slightly more affected (10,17). Clinical presentation of EECC includes symptoms such as otorrhea, otalgia, hearing loss, ear fullness and chronic itching ear (14). Some cases can be asymptomatic, and others may present with complication such as facial palsy (8), temporomandibular joint pain (18), labyrinthine fistula, meningitis or intracranial abscess (19). In a meta-analysis, Dubach reported that the cardinal symptoms are unilateral otorrhea and unilateral otalgia, and that

hearing loss is not typical complaint for EECC (14). In our case, hearing loss was the main complaint, and it could be due to piston prothesis dislocation. Nevertheless, in Lin's study (20) and in Udayabhanu HN's study (17), hearing loss was more common in secondary EACCs. Physical examination usually shows occlusion of the ear canal with white keratin flakes and otorrhea accompanied by focal bony erosion in the osseous cartilaginous ear canal (21). The Localization in the floor of the external ear canal is highly typical of primary EECC, whereas the secondary EECC's localization is ubiquitous (21). Aural polyps may be seen, and in some advanced cases, there may be destruction of the posterior canal wall with open mastoid cells and detached bone sequestra (8,14). The tympanic membrane can be intact or present a small perforation (21). The audiological assessment is based on pure-tone audiometry. According to the study by He et al. almost 75% of patients have conductive or mixed hearing loss (10). The mean air conduction and air-bone gap averaged thresholds were 45.4 ± 17.9 dB and 24.6 ± 15 respectively (10). Radiological investigations are necessary confirm the diagnosis all forms of cholesteatoma, to assess its extension, and to plan the management (16). High resolution Computed Tomography (HRCT) is the gold standard in the evaluation of EECC (22). The presence of soft tissue hypodensity in the external ear canal with smooth or irregular surrounding bony erosion is suggestive of EECC (8). The hypodensity can be lining the wall of the ear canal or completely occluding the lumen of the canal (8). It may also contain bone fragments inside (13). Primary EACC shows typically erosion of the posterior and inferior canal walls, but according to expansion, there may be erosion of the anterior and superior canal walls or circumferential erosion (8). In the case of secondary EACC, erosion may be multifocal (17). HRCT is also valuable to evaluate the extension of the disease to the middle-ear cavity, mastoid, temporomandibular joint, dehiscence of the facial canal, labyrinth, semicircular canal, tegmen tympani and
sinus plate, because these features may change the surgical management (8). If there is any doubt, Diffusion-weighted magnetic resonance imaging can be helpful to confirm cholesteatoma diagnosis.

The main differential diagnosis of EECC includes keratosis obturans, osteoma, malignant tumor of EAC (Squamous cell carcinoma), necrotizing otitis external, postinflammatory medial canal fibrosis and late complication of a Langerhans cell histiocytosis (13,16,17).

Several staging systems for EECCs were proposed. The first one was published in 1992 by Holt who defined 3 EECC stages on macroscopic studies : I-Superficial, saucerized defect; small localized pit, II-Localized ear canal pocket, III- Extension into the mastoid (15). This staging does not include lesions beyond the temporal bone. Naim et al proposed in 2005 a staging system based on histopathological findings (23), but intra-operative sampling errors may be responsible of misjudgment between Stage II and III lesions (10). Shin et al. published in 2010 a staging system for EECC of 4 stages, based on clinical and radiological findings : I- EECC is limited to the external ear canal, II- Invasion of the tympanic membrane and middle ear in addition to external ear canal, III- EECC creates a defect in the external ear canal and involves the air cells in the mastoid bone, IV- Involves lesions beyond the temporal bone (12). Shin et al. provided management strategy for each stage (12). We used this staging system because it is easy to follow and helps to decide treatment approach. The latest staging system was proposed by He et al. in 2021, in which they added a stage without bony erosion (Stage I) and subdivided Stage III in 3 subtypes depending on middle ear and mastoid cells invasion (Table 1). Based on this staging system, our case can be classified Stage IIIA.

Stage	*Features*	*Treatment*
I	Invasion without bony lesions	Conservative treatment and serial endoscopic debridement
II	Invasion confined within the EAC, and possible bone erosion manifested as a rough edge or localized defect of the bone	Serial endoscopic debridement or Canaloplasty
III	Invasion beyond the EAC, involving the mastoid air cells or tympanic cavity, but confined within the temporal bone	
A	Backward invasion into the mastoid air cells	Mastoidectomy or partial mastoidectomy, and canaloplasty
B	Inward or upward invasion into the tympanic cavity	Canaloplasty and tympanoplasty
C	Invasion into the tympanic cavity and mastoid air cells	Mastoidectomy, Canaloplasty and tympanoplasty and reconstruction of the EAC
IV	Invasion beyond the temporal bone or complications caused by the involvement of structures adjacent to the temporal bone	Surgical technique is determined according to the extent of invasion

***Table 1** : Definition for the stages of external auditory canal cholesteatoma and the proposed treatment according de He et al* (10).

Management of EECCs is debated and depends on the extent of the disease and whether it is categorized primary or secondary (11). The treatment's goals are to remove totally the cholesteatoma, to recreate a normal functioning EAC with uninterrupted skin, self-cleaning proprieties in order to restore normal epithelial migration, and prevent recurrence (17,20). If EECC is localized, it can be managed efficiently with conservative approach (11). It consists of several debridement of keratin debris and sequestered bone, and frequent cleanings in the office under local anesthesia (19,23). Placing a piece of gauze moistened with antibiotic and hydrocortisone ointment is

recommended (6,23). Surgical removal of disease is required if the conservative treatment is not sufficient to control the clinical symptoms, or if the EECC is already in advanced stages with destruction of the bony ear canal and invasion of adjacent structures (19). The aim of surgery is therefore to completely remove the cholesteatoma, eroded skin and destroyed bone. The type of required surgical intervention depends on the extension of the disease and the surgeon decision (12). In general, cholesteatoma and all affected skin are removed leaving healthy skin edges only (7). Then the meatal skin can be raised, and the eroded bone must be removed. Then the canaloplasty may be done. The irregular bony surface should be saucerized with diamond burr in order to obtain a smooth surface (19). The bony defect can be covered pedicled soft-tissue periosteal flap, cartilage, perichondrium and temporalis fascia (16,19). If there is middle ear or mastoid involvement, partial mastoidectomy or modified radical mastoidectomy with or without tympanoplasty are required (10,12). He et al. suggested that it is not necessary to remove all the air cells, but only the involved air cells (10). Canal wall down technique or subtotal petrosectomy are also possible surgical options (17,23). If EECC extends beyond the temporal bone approaches such as the middle fossa approach or the transzygomatic approach can be used to control the disease (16). Naim et al. recommended histopathological examination of all removed EECC to rule out malignant tumors of the EAC (23). Regular follow up is necessary to remove crust and to prevent reaccumulation of keratin (16). Recurrence is possible and justifies long term follow up (12).

IV. Conclusion

External ear canal cholesteatoma is a rare entity in comparison to middle ear cholesteatoma. It is divided into primary or spontaneous EECC and secondary. Patients of any age and any gender can be affected. The main symptoms are otorrhea, otalgia and hearing loss, but they are nonspecific. Diagnosis is suspected on clinical examination. The presence of ear canal occlusion with keratin debris, focal skin disruption and bony erosion usually lead to disgnosis. Temporal bone HRCT is mandatory to differentiate EECC from other diseases of ear canal, to assess its extension and to plan the management strategy. Although primary localized EECC can be managed conservatively, surgery remain the treatment of choice to eradicate the lesion and to reconstruct the ear canal. The particularity of our case was the incidental discovery of the secondary postoperative EECC. The patient was completely asymptomatic prior to the car accident, which resulted in the dislocation of the piston prosthesis responsible for the hearing loss and tinnitus. The EECC removal, the canaloplasty and the piston prothesis repositioning were successfully performed by a postauricular approach.

Conflitcs Of Interest:
The authors have no conflict of interests to declare.